Thymoma with Paraneoplastic Syndromes, Good's Syndrome, and Pure Red Cell Aplasia

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Case Report

A 56-year-old man was examined for a 1.5-year history of recurrent airway infections and sinusitis. He had also been diagnosed with pure red cell aplasia (PRCA), hemoglobin concentration 4.7 mmol/l, for which he was treated with corticosteroids (prednisone 1 mg/kg/day) for 1 year and blood transfusions.[1] This treatment stabilized his hemoglobin concentration at 9.5 mmol/l. The recurrent airway infections were first thought to be treatment-related complications by corticosteroids and were treated with antibiotics. Sputum cultures grew *Haemophilus influenzae*. A computed tomographic scan to exclude bronchiectasis revealed a non-invasive thymoma measuring 11 × 5 cm (Figure 1).

Because the sinopulmonary infections did not diminish, additional tests were performed to exclude an immunodeficiency that can occur in the presence of a thymoma. The immunoglobulin levels were very low (IgG 3.2 g/l; IgA and IgM non-detectable), and the patient was diagnosed with Good's syndrome.[2] Treatment with intravenous gamma globulin was started every 6 weeks, which increased the IgG level to 10.2 g/l and led to less frequent pulmonary infections. The noninvasive and encapsulated thymoma was radically resected by median sternotomy[3] (Figure 2). Postoperative recovery was uneventful. Histology confirmed the diagnosis of thymoma, type A/B. As expected, the hypogammaglobulinaemia did not diminish. Therefore, intravenous gamma globulin therapy was continued every 6 weeks. The hematopoiesis, however, completely recovered after thymectomy.

Thymomas are associated with different paraneoplastic syndromes; myasthenia gravis, Lambert-Eaton myasthenic syndrome, pemphigus, subacute sensory neuronopathy, pure red cell aplasia, and immunodeficiency. Most of these are autoimmune or endocrine-related. The most clinically important are myasthenia gravis, PRCA, and hypogammaglobulinaemia. In the presence of a thymoma, PRCA occurs in 5% to 15% and hypogammaglobulinaemia in less than 5% of the cases. Treatment of PRCA consists of thymectomy. However, this may result in normalization of the bone marrow in only 30% to 40%. A recent case series of 12 patients with thymoma and PRCA reported no remission of anemia after thymectomy.[4] In contrast to other paraneoplastic disorders, hypogammaglobulinaemia is not influenced by thymectomy.

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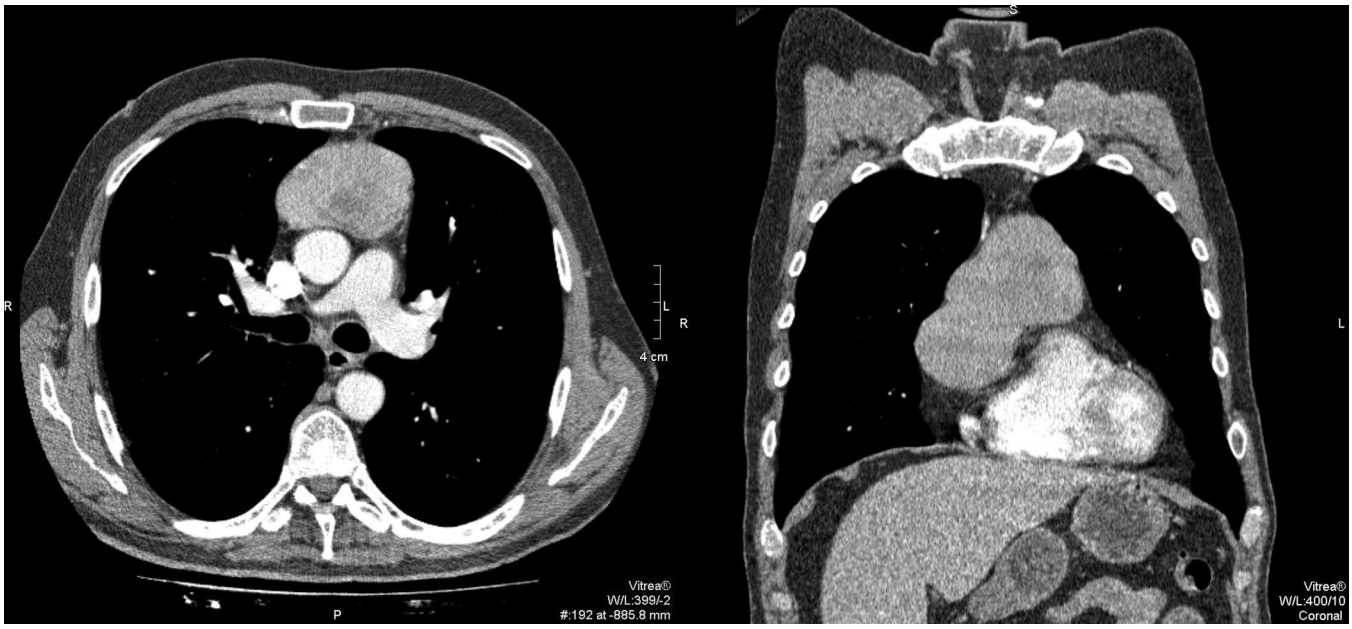

FIGURE 1. Chest computed tomographic scan showing a large mediastinal mass (thymoma). *A,* Axial view. *B,* Coronal view.

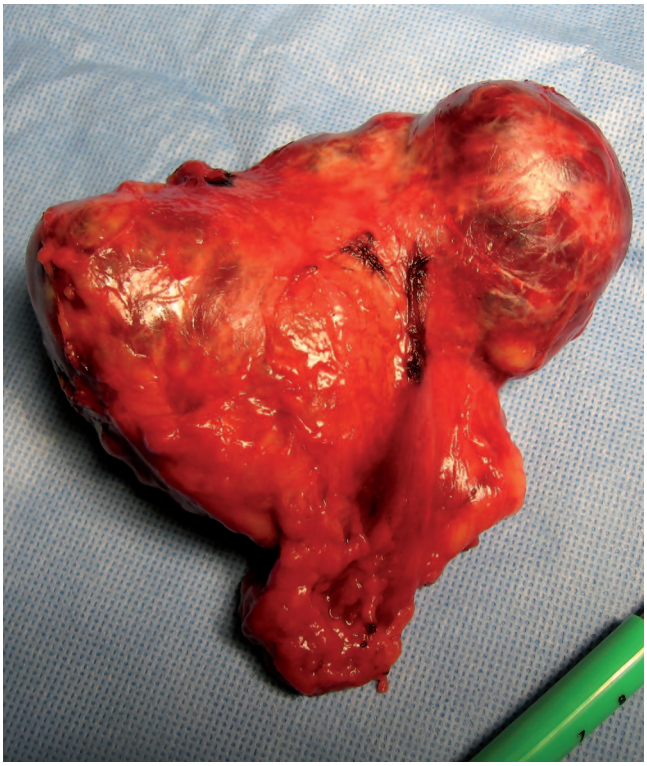

FIGURE 2. Radical resected thymoma. Type A/B, mixed thymoma. Size 11 × 5 cm.